



## Lennox-Gastaut syndrome

Lennox-Gastaut syndrome is a form of severe epilepsy that begins in childhood. It is characterized by multiple types of seizures and intellectual disability.

People with Lennox-Gastaut syndrome begin having frequent seizures in early childhood, usually between ages 3 and 5. More than three-quarters of affected individuals have tonic seizures, which cause the muscles to stiffen (contract) uncontrollably. These seizures occur most often during sleep. Also common are atypical absence seizures, which cause a partial or complete loss of consciousness. Additionally, many affected individuals have drop attacks, which are sudden episodes of weak muscle tone. Drop attacks can result in falls that cause serious or life-threatening injuries. Other types of seizures have been reported less frequently in people with Lennox-Gastaut syndrome.

Most of the seizures associated with Lennox-Gastaut syndrome are very brief. However, more than two-thirds of affected individuals experience at least one prolonged period of seizure activity known as nonconvulsive status epilepticus. These episodes can cause confusion and a loss of alertness lasting from hours to weeks.

Almost all children with Lennox-Gastaut syndrome develop learning problems and intellectual disability associated with their frequent seizures. Because the seizures associated with this condition are difficult to control with medication, the intellectual disability tends to worsen with time. Some affected children develop additional neurological abnormalities and behavioral problems. Many also have delayed development of motor skills such as sitting and crawling. As a result of their seizures and progressive intellectual disability, most people with Lennox-Gastaut syndrome require help with some or all of the usual activities of daily living. However, a small percentage of affected adults live independently.

People with Lennox-Gastaut syndrome have an increased risk of death compared to their peers of the same age. Although the increased risk is not fully understood, it is partly due to poorly controlled seizures and injuries from falls.

### Frequency

Lennox-Gastaut syndrome affects an estimated 1 in 50,000 to 1 in 100,000 children. This condition accounts for about 4 percent of all cases of childhood epilepsy. For unknown reasons, it appears to be more common in males than in females.

## **Genetic Changes**

Researchers have not identified any genes specific to Lennox-Gastaut syndrome, although the disorder likely has a genetic component. About two-thirds of cases are described as symptomatic, which means that they are related to an existing neurological problem. Symptomatic Lennox-Gastaut syndrome can be associated with brain injuries that occur before or during birth, problems with blood flow in the developing brain, brain infections, or other disorders affecting the nervous system. The condition can also result from a brain malformation known as cortical dysplasia or occur as part of a genetic disorder called tuberous sclerosis complex. Many people with Lennox-Gastaut syndrome have a history of recurrent seizures beginning in infancy (infantile spasms) or a related condition called West syndrome.

In about one-third of cases, the cause of Lennox-Gastaut syndrome is unknown. When the disorder occurs without an apparent underlying reason, it is described as cryptogenic. Individuals with cryptogenic Lennox-Gastaut syndrome have no history of epilepsy, neurological problems, or delayed development prior to the onset of the disorder.

## **Inheritance Pattern**

Most cases of Lennox-Gastaut syndrome are sporadic, which means they occur in people with no history of the disorder in their family. However, 3 to 30 percent of people with this condition have a family history of some type of epilepsy. People with the cryptogenic form of Lennox-Gastaut syndrome are more likely than people with the symptomatic form to have a family history of epilepsy.

## **Other Names for This Condition**

- childhood epileptic encephalopathy with diffuse slow spikes and waves
- LGS

## **Diagnosis & Management**

### Genetic Testing

- Genetic Testing Registry: Epileptic encephalopathy Lennox-Gastaut type  
<https://www.ncbi.nlm.nih.gov/gtr/conditions/C1853372/>

### Other Diagnosis and Management Resources

- Cleveland Clinic  
<http://my.clevelandclinic.org/childrens-hospital/health-info/diseases-conditions/hic-Lennox-Gastaut-Syndrome>
- National Institute of Neurological Disorders and Stroke: Diagnosis and Treatment of Epilepsy  
<https://www.ninds.nih.gov/Disorders/All-Disorders/Epilepsy-Information-Page#158133109>
- News Release: FDA Approves New Drug to Treat Severe Form of Epilepsy (U.S. Food and Drug Administration, November 20, 2008)  
<https://www.fda.gov/NewsEvents/Newsroom/PressAnnouncements/2008/ucm116980.htm>

### General Information from MedlinePlus

- Diagnostic Tests  
<https://medlineplus.gov/diagnostictests.html>
- Drug Therapy  
<https://medlineplus.gov/drugtherapy.html>
- Genetic Counseling  
<https://medlineplus.gov/geneticcounseling.html>
- Palliative Care  
<https://medlineplus.gov/palliativecare.html>
- Surgery and Rehabilitation  
<https://medlineplus.gov/surgeryandrehabilitation.html>

### **Additional Information & Resources**

#### MedlinePlus

- Encyclopedia: Seizures  
<https://medlineplus.gov/ency/article/003200.htm>
- Health Topic: Epilepsy  
<https://medlineplus.gov/epilepsy.html>

#### Genetic and Rare Diseases Information Center

- Lennox-Gastaut syndrome  
<https://rarediseases.info.nih.gov/diseases/9912/lennox-gastaut-syndrome>

### Additional NIH Resources

- National Institute of Neurological Disorders and Stroke  
<https://www.ninds.nih.gov/Disorders/All-Disorders/Lennox-Gastaut-Syndrome-Information-Page>

### Educational Resources

- AboutKidsHealth: The Hospital for Sick Children (Canada)  
<http://www.aboutkidshealth.ca/En/ResourceCentres/Epilepsy/UnderstandingEpilepsyDiagnosis/EpilepsySyndromes/Pages/Lennox-Gastaut-Syndrome.aspx>
- Cleveland Clinic  
<http://my.clevelandclinic.org/childrens-hospital/health-info/diseases-conditions/hic-Lennox-Gastaut-Syndrome>
- MalaCards: lennox-gastaut syndrome  
[http://www.malacards.org/card/lennox\\_gastaut\\_syndrome](http://www.malacards.org/card/lennox_gastaut_syndrome)
- Orphanet: Lennox-Gastaut syndrome  
[http://www.orpha.net/consor/cgi-bin/OC\\_Exp.php?Lng=EN&Expert=2382](http://www.orpha.net/consor/cgi-bin/OC_Exp.php?Lng=EN&Expert=2382)
- Washington University School of Medicine  
<https://neuro.wustl.edu/patient-care/pediatric-neurology/pediatric-epilepsy-center/patient-family-and-physician-information/lennox-gastaut-syndrome/>

### Patient Support and Advocacy Resources

- National Organization for Rare Disorders (NORD)  
<https://rarediseases.org/rare-diseases/lennox-gastaut-syndrome/>
- Tuberous Sclerosis Alliance  
<http://www.tsalliance.org/>

### ClinicalTrials.gov

- ClinicalTrials.gov  
<https://clinicaltrials.gov/ct2/results?cond=%22Lennox-Gastaut+syndrome%22>

### Scientific Articles on PubMed

- PubMed  
<https://www.ncbi.nlm.nih.gov/pubmed?term=%28Epilepsy%5BMAJR%5D%29+AND+%28lennox-gastaut+syndrome%5BTIAB%5D%29+AND+english%5Bla%5D+AND+human%5Bmh%5D+AND+%22last+1800+days%22%5Bdp%5D>

### OMIM

- MACROCEPHALY AND EPILEPTIC ENCEPHALOPATHY  
<http://omim.org/entry/606369>

## Sources for This Summary

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